

**COUNTY OF LOS ANGELES - DEPARTMENT OF HEALTH SERVICES  
PUBLIC HEALTH - DISEASE CONTROL PROGRAMS  
VETERINARY PUBLIC HEALTH AND RABIES CONTROL**

**SPONGIFORM ENCEPHALOPATHIES**

Transmissible spongiform encephalopathies are also known as prion diseases because they are thought to be caused by abnormal forms of a host protein, termed prion protein (PrP). These agents cause transmissible spongiform encephalopathies, including: scrapie of sheep, kuru (associated with human cannibalism), Creutzfeldt-Jakob disease (CJD associated with corneal transplants), bovine spongiform encephalopathy (BSE), better known as mad cow disease.

Background information on BSE is available online

[http://www.avma.org/pubhlth/bse/bse\\_bgnd.asp](http://www.avma.org/pubhlth/bse/bse_bgnd.asp). A variant Creutzfeldt-Jakob disease (vCJD), is likely transmitted to humans from BSE-infected cattle. Newly recognized transmissible spongiform encephalopathies seen in captive wild ruminants, domestic cats, and captive wild cats are likely related to consumption of prion infected tissue.

Diseases occur when the prion protein undergoes a conformational change that confers resistance to proteases. The incubation period in prion diseases is measured in terms of years. The spongiform encephalopathy of sheep, commonly known as scrapie, is the unofficial prototype for prion diseases. Veterinarians have struggled for over a century with scrapie.

**Scrapie**

Scrapie is a fatal, neurologic disease that produces subacute spongiform encephalopathy in adult sheep. Similar diseases occur naturally in goats. Scrapie is frequently transmitted in family lines in flocks, which indicates that some form of maternal transmission may occur at a pre- or postnatal stage. Infected placentas eaten by other sheep, or contaminate pastures, may account for horizontal transmission. During the first eight months following birth, infection is undetectable in any sheep tissue bioassayed in mice. Clinical signs in sheep typically start at 3½ years of age.

Scrapie has been endemic in Europe for more than 200 years and was first recognized in the United States in 1947 with sheep imported into Michigan. Los Angeles County's last documented outbreak occurred in 1990. The sheep in that flock were destroyed.

**Transmissible mink encephalopathy**

This progressive neurologic disease of mink is rare, but the mortality rate may reach 60- 90% of the ranch population. The incubation period is about 12 months. Affected mink usually bite compulsively, are incoordinated and somnolent, scatter feces in the pen, and flip their tails up over their backs (like a squirrel). Histologic lesions of the brain are spongiform changes of the gray matter, astrogliosis, and neuronal vacuolation. The demonstration of disease-specific prion protein in nervous tissues aids in the diagnosis. While the means of transmission is unknown, it is highly likely that "downer" cattle are the source of the agent. There are no vaccines or treatment.

## **Kuru**

Kuru, a fatal spongiform encephalopathy in native people of the New Guinea highlands, was transmitted during cannibalistic practices when the brains of dead relatives were eaten. Missionaries discouraged the practice before it was known that the disease was transmissible in the food chain. Cannibalism has been abandoned and kuru has disappeared. The longest, documented, incubation period in kuru is over thirty years.

## **Chronic Wasting Disease**

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy of deer and elk. To date, this disease has been found only in cervids (members of the deer family). First recognized as a clinical "wasting" syndrome in 1967 in mule deer in a wildlife research facility in northern Colorado, it was identified as a transmissible spongiform encephalopathy in 1978. CWD is typified by chronic weight loss, leading to death. There is no known relationship between CWD and any other TSE of animals or people.

In the mid-1980s, CWD was detected in free-ranging deer and elk in contiguous portions of northeastern Colorado and southeastern Wyoming. In May 2001, CWD was also found in free-ranging deer in the southwestern corner of Nebraska (adjacent to Colorado and Wyoming) and later, in additional areas in western Nebraska. The limited area of northern Colorado, southern Wyoming, and western Nebraska in which free-ranging deer and/or elk positive for CWD have been found, is referred to as the endemic area.

CWD also has been diagnosed in farmed elk and deer herds in a number of States and in two Canadian provinces. The first positive farmed elk herd in the United States was detected in 1997 in South Dakota. The Texas Parks and Wildlife Department has a website on the disease [http://www.tpwd.state.tx.us/hunt/chronic\\_wasting\\_disease/](http://www.tpwd.state.tx.us/hunt/chronic_wasting_disease/).

A study was published in *Emerging Infectious Diseases* Vol 9, No5 May 2003 on "Chronic Wasting Disease in Free-Ranging Wisconsin White-Tailed Deer" <http://www.cdc.gov/ncidod/EID/vol9no5/02-0721.htm>. The study investigated three White-tailed Deer shot within 5 km of each other, during the 2001 hunting season in Wisconsin which tested positive for chronic wasting disease. Subsequent sampling within 18 km, showed a 3% prevalence. This discovery represents an important range extension for chronic wasting disease into the eastern United States.

**General reference:** Kumar: Robbins and Cotran: *Pathologic Basis of Disease*, 7th edition 2005 pages 346, 1380-82

2005

Spongiform encephalopathies05a.doc